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Fibrous Dysplasia of the Temporal Bone: Reversal of Sensorineural Hearing Loss After Decompression of the Internal Auditory Canal

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When fibrous dysplasia affects the temporal bone, it most often presents with conductive hearing loss attributable to stenosis of the external auditory canal. Sensorineural hearing loss has usually been attributed to involvement of the otic capsule. We present a patient with bilateral fibrous dysplasia of the temporal bones who complained of unilateral hearing loss, facial tingling, and facial twitching. The audiogram showed severe sensorineural hearing loss. The hearing markedly improved and facial twitching and tingling ceased after decompression of the internal auditory canal via a middle fossa approach. This is the only case of which we are aware showing reversal of sensorineural hearing loss caused by fibrous dysplasia.

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INTRODUCTION

Fibrous dysplasia is a bone disorder of unknown origin in which normal bone is replaced with fibrotic tissue and disorganized bonetrabeculae. It was first described by McCune and Bruch in 1937. The disease has since been found to have three different variants: monostotic, polyostotic, and Albright's syndrome. This disorder occasionally affects the temporal bone. Most involvement of the temporal bone is unilateral and is associated with conductive hearing loss attributable to compression of the external auditory canal. Sensorineural loss usually is due to otic capsule involvement by fibrous dysplasia or secondary cholesteatoma.[1] Two other cases with bilateral temporal bone involvement have been reported.[2] We present a patient with bilateral fibrous dysplasia of the temporal bones associated with unilateral sensorineural hearing loss, facial twitching, and facial tingling.

CASE REPORT

A 28-year-old white woman (height, 1.7 m; weight, 113 kg) presented with slowly progressive hearing loss over a 2-year period and the recent onset of facial twitching. She also complained of mild, continuous left-side tinnitus and a sensation of facial tingling as if her face was "a little asleep." She denied any vestibular symptoms or facial weakness. She had no history of otologic surgery. Review of systems revealed a sense of tongue irritation and "funny taste." She had originally been diagnosed with fibrous dysplasia at the age of 10 years when a 6 × 6 cm mass of fibrous dysplasia was curretted from her occipital bone. When she was 11 years old, surgeons removed another golf ball–sized mass from her occipital region. Since that time she had no other difficulties associated with the disease. Her medical history included pseudocholinesterase deficiency, insulin-dependent diabetes, and hypothyroidism. The onset of the diabetes was during her first pregnancy. The hypothyroidism, diagnosed during her second pregnancy, was due to pituitary insufficiency and not clearly pregnancy related or congenital. Other surgeries include tonsillectomy and adenoidectomy at age 22 years and hysterectomy at age 23 years attributable to menorrhagia. On physical examination her external auditory canals and tympanic membranes appeared normal. She had no visible or palpable abnormalities of the skull. The remainder of her physical examination was unremarkable. The audiogram showed a left-side sensorineural hearing loss with a pure-tone average of 70 dB and a speech reception threshold of 75 dB. Hearing on the right side was normal. There was no family history of bone disease, skin disorders, early

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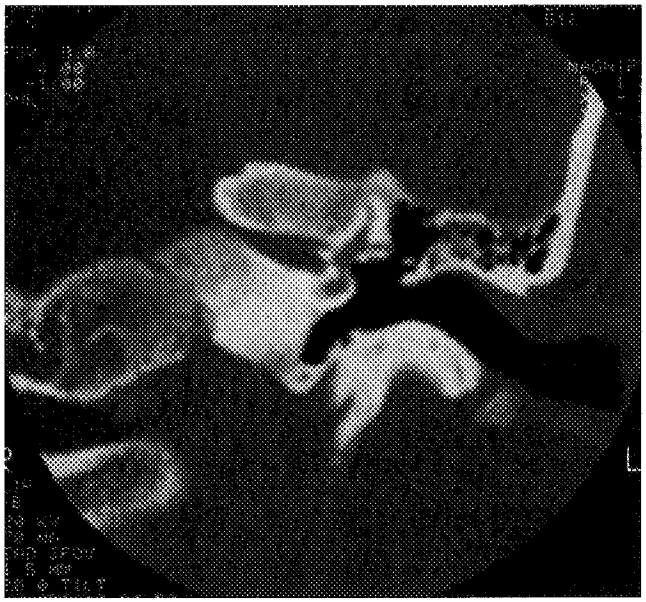

Fig. 1. Coronal computed tomographic image of the left-side temporal bone showing severe narrowing of the porus of the internal auditory canal. There is no involvement of the otic capsule, middle ear, or external auditory canal.

puberty in the women, or other endocrine or genetic abnormalities. Serum calcium, phosphorus, and alkaline phosphatase levels were in the normal range.

Computed tomography of the temporal bone showed extensive thickening of the diploic and medullary spaces with ground glass–appearing, moderately dense new bone and fibrous tissue. There was considerable expansion of the petrous apex and narrowing of the internal auditory canal. The squamous portion of the temporal bone was spared, which is typical of fibrous dysplasia of the temporal bone. The middle ear, ossicular chain, and facial nerve canal all appeared normal (Figs. 1 and 2). The dense bone of the cochlear otic capsule showed some thinning. There was milder involvement of the right petrous apex with some narrowing of the internal auditory canal (Fig. 3). The clivus demonstrated a predominately "lytic" or radiolucent process reflecting the greater fibrous content of the lesion in that region. Both the inner and outer cortices of the involved bones were intact, an important characteristic of fibrous dysplasia. This case is somewhat unusual in that the petrous apices are more involved than the mastoid and external auditory canals.

The patient underwent decompression of the left internal auditory canal and debulking of the expansile mass through a middle fossa approach. Decadron was given intraoperatively and tapered over 3 days. She tolerated the procedure well with mild headaches and intermittent unsteadiness, which resolved within 8 weeks. Follow-up audiogram showed improvement to 10 dB pure-tone average, speech reception threshold of 5 dB, and speech discrimination of 100% (Fig. 4). Her facial twitching and abnormal taste sensation resolved. She had no return of symptoms with 3 years of follow-up.

DISCUSSION

Historically, fibrous dysplasia was grouped with other bone diseases such as Paget's disease and von Recklinghausen's disease.[1] Recognition as a separate disease began in 1937 when McCune reported the case of a young girl described as having osteodystrophia fibrosa, pigmented skin lesions, and precocious puberty.[1] Later that year five similar cases were described by Albright et al.[3] Subsequently this variant of fibrous dysplasia became known as Albright's syndrome. The term "fibrous dysplasia" was suggested by Lichtenstein in 1938.[4]

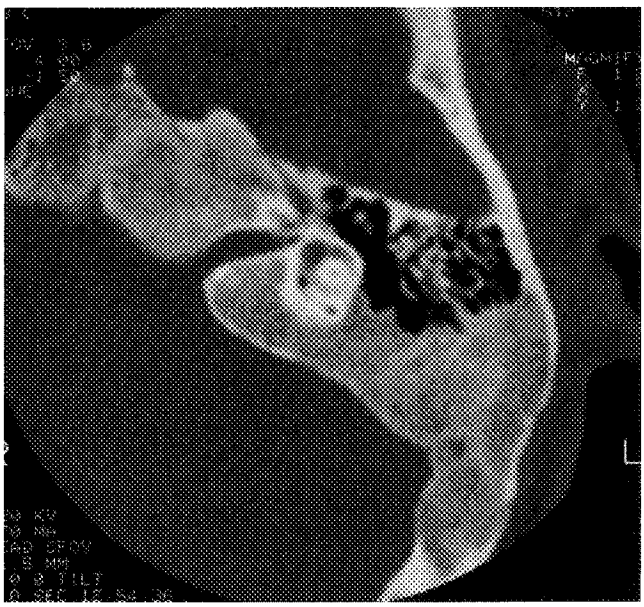

Fig. 2. Axial computed tomogram of the left temporal bone. Fibrous dysplasia has caused expansion of the petrous apex. The overlying cortical bone is intact.

Fig. 3. Axial computed tomogram of the right temporal bone. There is expansion of the petrous apex and narrowing of the porus of the internal auditory canal resulting from fibrous dysplasia.

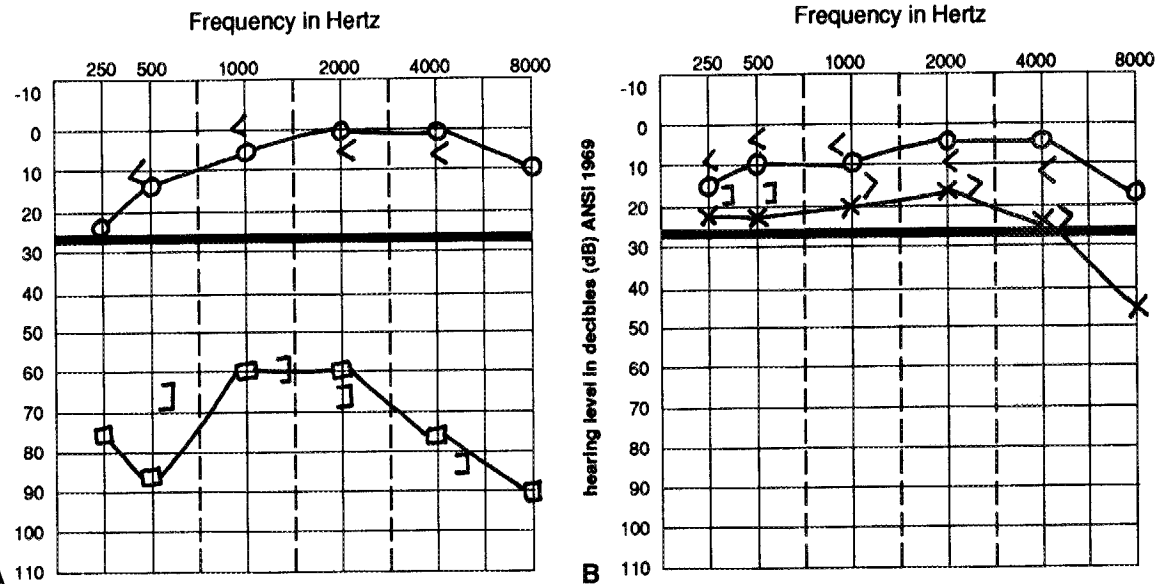

Fig. 4. Audiograms before **(A)** and 3 years after surgery **(B)** showing reversal of severe left-sided sensorineural hearing loss.

Fibrous dysplasia is a benign disease in which there is slowly progressive growth of abnormal fibroosseous tissue which displaces the normal medullary bone. The tissue contains disorganized bony trabeculae and spindle cells surrounded by a fibrous matrix.[2,5–7] Theories of etiology for this disease include aberrant differentiation of mesenchyme during bone formation, an arrest of bone at the immature woven stage, or a disturbance of cancellous bone maintenance.[8] The tissue expands, causing distortion and weakening of the bone. The overlying cortical bone may be thinned because of pressure but is not directly involved in the disease process.[5,8]

Three variants of fibrous dysplasia are recognized. Monostotic fibrous dysplasia involves a single bone, polyostotic fibrous dysplasia involves two or more bones, and Albright's syndrome is polyostotic disease associated with pigmented skin lesions, endocrine abnormalities, and precocious puberty.

Monostotic fibrous dysplasia accounts for 70% of cases and tends to become evident in late childhood. The lesions usually become quiescent after puberty.

Polyostotic fibrous dysplasia accounts for about 30% of cases and usually becomes evident earlier as a result of multiple bone involvement. It is more likely to have ongoing progression of the lesions into the third and forth decades. New lesions may also develop during adulthood. It also differs from the monostotic form of the disease in that it more commonly affects the shoulder, pelvis, vertebral column, and craniofacial skeleton. It is more likely to affect entire limbs and cause severe crippling deformities and spontaneous and recurrent fractures.[8]

Albright's syndrome is a variant of polyostotic fibrous dysplasia associated with areas of abnormal skin pigmentation (usually unilateral on the side of the body with more bone lesions), endocrine abnormalities, and precocious puberty. It is seen almost exclusively in females; however, a few cases of the disease in males have been reported.[3,8,9] Presentation is earliest in this variant because of the associated endocrine abnormalities. The skeletal lesions are polyostotic, typically with a unilateral distribution. The areas of increased skin pigmentation may be large and usually are found on the neck, back, chest, shoulder, and pelvic girdle. The endocrine abnormalities include hyperthyroidism with goiter, acromegaly, Cushing's disease, hyperparathyroidism, and extrainsular hypothalamic diabetes mellitus. Hyperthyroidism is the most common associated endocrinopathy, being seen in 5% of patients with Albright's syndrome.[10]

Malignant transformation is seen in approximately 0.5% of patients.[11–13] Patients with Albright's syndrome have a reported incidence of 4%.[14] The average time between diagnosis of fibrous dysplasia and malignancy is 13.5 years. The malignancies reported have included osteosarcoma, fibrosarcoma, chondrosarcoma, and giant cell sarcoma. Malignant transformation has never been reported in temporal bone disease.[15] Past experience showed an increase in the incidence of malignant transformation to 44% after radiation treatment.[13]

Because of the slow progression of the disease, it may be years before the patient becomes symptomatic. Many mild cases may never be recognized or are found incidentally. Craniofacial involvement is seen in 10% to 30% of cases of monostotic disease and about 50% to 100% of cases of polyostotic disease depending on the severity.[8] In a review of 144 patients with skull lesions caused by fibrous dyspla-

sia, Van Tilburg[16] found the temporal bone to be affected 18% of the time when the skull was involved.[2]

Abnormal skin pigmentation is the most common of the extraskeletal manifestations of the disease. It is occasionally found in the monostotic form of the disease. Polyostotic disease is associated with more than 50% incidence of skin lesions; in Albright's syndrome it is an almost universal finding.

Hearing loss and visual impairment are the most common neurologic symptoms of fibrous dysplasia involving the craniofacial skeleton.[6] Visual symptoms are due to compression of the optic nerve and can lead to permanent blindness. Hearing loss is most often conductive as a result of stenosis of the external auditory canal. Cholesteatoma is seen in 40% of cases and usually originates from the external canal wall. Sensorineural loss attributed to the lesion is seen in 17% of patients with fibrous dysplasia of the temporal bone.[2] Facial nerve involvement is seen in 10% of these patients. Although not common, function of all cranial nerves can be affected in cases of fibrous dysplasia.[8]

Laboratory studies should include evaluation of serum calcium, phosphorus, and alkaline phosphatase levels. One series of 124 patients showed elevated levels of serum calcium between 11 to 13 mg/dL in 25% of patients.[17] In another study the alkaline phosphatase level was reported as elevated in 25% of patients with monostotic and 67% of patients with polyostotic fibrous dysplasia.[18] However, in a review of 69 patients by Nager et al.[8] every patient had normal calcium and phosphorus levels. A single patient had an abnormally elevated serum alkaline phosphatase level.[8]

On radiographic examination fibrous dysplasia has a characteristic homogenous ground-glass appearance and is usually surrounded by a shell of dense cortical bone. Expansion of the lesions can lead to bone remodeling. Craniofacial fibrous dysplasia may have several radiographic patterns: pagetoid, sclerotic, and cystic.[2,14,15] The pagetoid pattern seen in 56% of cases is typified by a mixture of dense and radiolucent areas of fibrosis with bone expansion. The sclerotic pattern is homogenously dense with bone expansion seen on 23% of radiographic studies. Spheric or oval lucent regions with a dense boundary are typical of the cystic pattern seen in 21% of cases. Temporal bone lesions have a sclerotic pattern in two thirds of cases. Patients may have more than one of these patterns on the radiographic studies. The radiographic differential diagnosis includes Paget's disease, ossifying fibroma, osteoma, meningioma, hemangioma, and mucocele. Paget's disease produces expansion of the bone similar to fibrous dysplasia. However, areas of cortical erosion and the characteristic coarse bony trabeculae of Paget's disease differentiate it from fibrous dysplasia. Ossifying fibroma is a true neoplasm and grows outward from a finite center producing mass effect on adjacent structures. This is in contrast to fibrous dysplasia, which produces a diffuse expansion of the medullary cavity. The radiographic density and histologic appearance of fibrous dysplasia and ossifying fibroma are similar, so the growth pattern is helpful in differentiating the two entities. Meningioma can induce bony thickening and sclerosis and may be confused with fibrous dysplasia. With meningioma there is always a soft tissue component adjacent to the bone; this is not seen in fibrous dysplasia. Hemangiomas may expand bone but usually contain very coarse, raylike heavy bone trabeculae. Mucocele might be confused with the purely radiolucent type of fibrous dysplasia that contains no bone spicules. Mucoceles have a well-defined bone margin and an expansile growth pattern, as opposed to the poorly marginated diffuse expansion of the medullary space seen in fibrous dysplasia. If necessary, magnetic resonance imaging (MRI) differentiates a mucocele, which contains mucous signal intensity and shows a lack of enhancement.

Treatment and definitive diagnosis rely on surgical debulking and biopsy. The decision to treat depends on the degree of cosmetic or functional impairment. Complications such as visual loss, hearing loss, and cholesteatoma are all surgical indications. Prognosis is good in most cases depending on the severity of disease. Use of periodic computed tomography can be helpful to follow the progression of disease and to assess the need for any surgical intervention.[15]

A review of 69 patients with fibrous dysplasia of the temporal bone was published by Nager et al.[8] The male-to-female ratio was 2:1. Race predilection was shown with whites comprising 80%, blacks 2%, and Asians 1% of cases. Race was not mentioned in all reports. The most common presenting symptoms were progressive hearing loss (56%), increasing size of the temporal bone (50%), and progressive occlusion of the external auditory canal (42%). Other symptoms included tinnitus, dizziness, pain, and trismus secondary to modeling of the glenoid fossa. Seven patients had total loss of cochlear function, one had profound sensorineural hearing loss, and 35 had conductive hearing loss attributable to external canal stenosis. Eleven patients had secondary cholesteatomas. Five had facial nerve palsy. One patient had cranial nerves II and V to X all affected; another had cranial nerves IV, V, and VI affected; yet another showed involvement of cranial nerves I to XII. Surgery was performed on 41 of the 69 patients; 20 required two or more surgeries. All patients had normal calcium levels. Only one patient showed an elevated serum alkaline phosphatase level.

Three cases of progression from conductive to profound sensorineural hearing loss were reported by Megerian et al.[2] All had involvement of the otic

capsule either by cholesteatoma or fibrous dysplasia. The researchers also reported a case of acute facial nerve paralysis that was decompressed from the geniculate ganglion to the lateral internal auditory canal with a functional return to House-Brackmann grade III; the patient had profound sensorineural hearing loss attributable to invasion of the cochlea. There is also a report of a patient with symptoms of sensorineural hearing loss and facial palsy caused by an expansile mass of fibrous dysplasia at the petrous apex. No mention was made of treatment or neurologic outcome.[14]

CONCLUSION

Fibrous dysplasia is a slowly progressive disease that can affect all bones of the body. There have been 76 cases of fibrous dysplasia affecting the temporal bone reported in the literature.[7] A total of nine cases of sensorineural hearing loss secondary to fibrous dysplasia have been reported.[2] None of these patients recovered their hearing. We report a case of a young woman with polyostotic fibrous dysplasia affecting both temporal bones with narrowing of the medial aspect of the internal auditory canal, causing unilateral involvement of cranial nerves VII and VIII. This is the only case of which we are aware that showed recovery of sensorineural hearing loss after surgical decompression.

This case demonstrates that surgical decompression may be of benefit in fibrous dysplasia and other disease of the temporal bone in which there is narrowing of the internal auditory canal. This may allow for preservation and possibly restoration of function of both the facial and vestibulocochlear nerves.

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EDITORIAL FOOTNOTE

During the review process it was noted that the patient received intraoperative and postoperative steroids. One reason for hearing improvement may be the procedure of internal auditory canal decompression, but it is also possible that the administration of steroids could have contributed to the reversal of the sensorineural hearing loss to some degree.

EDITOR